Appendices

Appendix A: Denominators Used in Calculating Rates

Live births in the area covered by the Texas Birth Defects Monitoring Division during 1996 and 1997, by mother's age, mother's race/ethnic group, infant's sex, and year and region.

		Number of Live Births
Overall		300,431
By Mother's age	<20	48,401
	20 - 24	83,398
	25 - 29	81,442
	30 - 34	57,562
	35+	29,574
	unknown	54
By Mother's race/ethnic group	White	102,193
	African American	29,254
	Hispanic	160,094
	Other	8,890
By Infant's sex	Male	153,534
	Female	146,897
By Year and Region	1996, Region 06	77,433
	1996, Region 11	36,651
	1997, Region 02	7,325
	1997, Region 03	85,469
	1997, Region 08	33,559
	1997, Region 09	8,331
	1997, Region 10	14,792
	1997, Region 11	36,871

Appendix B: BPA codes used to define the birth defects shown in this report

Diagnoses in the Texas Birth Defects Registry are coded using a system developed and provided by the National Center for Environmental Health of the Centers for Disease Control and Prevention (CDC). The birth defect codes, commonly called BPA codes, are based on the British Pediatric Association Classification of Diseases (1979) and the World Health Organization's International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM) (1979).

The table below shows the BPA codes used to define the conditions shown in this report.

Birth defect	BPA codes
Anencephaly	740.00 - 740.10
Spina bifida without anencephaly	741.00 - 741.99, without 740.00 - 740.10
Encephalocele	742.00 - 742.09
Microcephaly	742.10
Holoprosencephaly	742.26
Hydrocephaly	742.30 - 742.39, excluding 742.385
Anophthalmia	743.00
Microphthalmia	743.10
Cataract	743.32
Aniridia	743.42
Anotia or microtia	744.01, 744.21
Common truncus	745.00 - 745.01
Transposition of the great vessels	745.10 - 745.19
Tetralogy of Fallot	745.20 - 745.21, 746.84
Ventricular septal defect	745.40 - 745.49
Atrial septal defect	745.51 - 745.59
Endocardial cushion defect	745.60 - 745.69
Pulmonary valve atresia or stenosis	746.00 - 746.01
Tricuspid valve atresia or stenosis	746.10, excluding 746.105
Ebstein anomaly	746.20
Aortic valve stenosis	746.30
Hypoplastic left heart syndrome	746.70
Patent ductus arteriosus	747.00
Coarctation of the aorta	747.10 - 747.19
Choanal atresia or stenosis	748.00
Agenesis, aplasia, or hypoplasia of the lung	748.50 - 748.51
Cleft palate alone (without cleft lip)	749.00 - 749.09
Cleft lip with or without cleft palate	749.10 - 749.29
Tracheoesophageal fistula / esophageal atresia	750.30 - 750.35
Pyloric stenosis	750.51
Stenosis or atresia of small intestine	751.10 - 751.19
Stenosis or atresia of large intestine, rectum, or anal canal	751.20 - 751.24

Birth defect	BPA codes
Hirschsprung disease	751.30 - 751.34
Biliary atresia	751.65
Hypospadias or epispadias	752.60 - 752.62, excluding 752.621
Renal agenesis or dysgenesis	753.00 - 753.01
Obstructive genitourinary defect	753.20 - 753.29 and 753.60 - 753.69
Bladder exstrophy	753.50
Congenital hip dislocation	754.30
Reduction defects of the upper limbs	755.20 - 755.29
Reduction defects of the lower limbs	755.30 - 755.39
Craniosynostosis	756.00 - 756.03
Diaphragmatic hernia	756.61
Omphalocele	756.70
Gastroschisis	756.71
Down syndrome (includes trisomy 21, translocations, and mosaics)	758.00 - 758.09
Patau syndrome (trisomy 13)	758.10 - 758.19
Edwards syndrome (trisomy 18)	758.20 - 758.29, excluding 758.295
Fetal alcohol syndrome or other alcohol related birth defects	760.710, 760.719, 760.720, 760.729
Possible/probable FAS or other alcohol related birth defects	760.718, 760.728

Appendix C: Glossary of Birth Defect Terms

Agenesis Absence of part(s) of the body.

Agenesis, aplasia, or hypoplasia of the lung The absence or incomplete development of a lung or lung tissue.

Anencephaly Congenital absence of the skull, with cerebral hemispheres completely missing or reduced to small masses attached to the base of the skull. Anencephaly is not compatible with life.

Aniridia The complete absence of the iris of the eye or a defect of the iris. Can be congenital or traumatically induced.

Anopthalmia A developmental defect characterized by complete absence of the eyes, or by the presence of vestigial eyes.

Anotia A congenital absence of one or both ears.

Aortic valve stenosis A cardiac anomaly characterized by a narrowing or stricture of the aortic valve. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can be repaired surgically in some cases.

Atresia Imperforation; absence or closure of a normal opening.

Atrial septal defect A congenital cardiac malformation in which there are one or several openings in the atrial septum (muscular and fibrous wall between the right and left atria) allowing a mixing of oxygenated and unoxygenated blood. The openings vary in size and may resolve without treatment or may require surgical treatment. Also called *ostium secundum defect.*

Biliary atresia A congenital absence or underdevelopment of one or more of the ducts in the biliary tract. Correctable surgically.

Bladder exstrophy Incomplete closure of the anterior wall of the bladder and the abdominal cavity. The upper urinary tract is generally normal. Often associated with anorectal and genital malformations, and epispadias. Affected persons are at a markedly increased risk of bladder carcinoma (squamous cell). This condition is usually corrected surgically after birth.

Cataract An opacity (clouding) of the lens of the eye.

Choanal atresia or stenosis A congenital anomaly in which a bony or membranous formation blocks the passageway between the nose and the pharynx. This defect is usually repaired surgically after birth. Bilateral choanal atresia is a surgical emergency.

Cleft lip The congenital failure of the fetal components of the lip to fuse or join, forming a groove or fissure in the lip. Infants with this condition can have difficulty feeding, and may use assistive devices for feeding. This condition is corrected when the infant can tolerate surgery.

Cleft palate The congenital failure of the palate to fuse properly, forming a grooved depression or fissure in the roof of the mouth. This defect varies in degree of severity. The fissure can extend into the hard and soft palate and into the nasal cavities. Infants with this condition have difficulty feeding, and may use assistive devices for feeding. Surgical correction is begun as soon as possible. Children with cleft palates are at high risk for hearing problems due to ear infections.

Coarctation of the aorta Localized narrowing of the aorta. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can vary from mild to severe. Surgical correction is recommended even for mild defects.

Common truncus ateriosus A congenital heart defect in which the common arterial trunk fails to divide into pulmonary artery and aorta. This is corrected surgically.

Confidence interval (95%) The interval that contains the true prevalence (which we can only estimate) 95% of the time. See Methods for more explanation.

Congenital Existing at or dating from birth.

Congenital hip dislocation A congenital defect in which the head of the femur does not articulate with the acetabulum of the pelvis because of an abnormal shallowness of the acetabulum. Treatment in early infancy consists of bracing of the joint to cause a deepening of the acetabulum.

Craniosynostosis A premature ossification (closing) of the cranial sutures before birth or soon after birth. This condition is occasionally associated with other skeletal defects. If no surgical correction is made, the growth of the skull is inhibited, and the head is deformed. The eyes and the brain are often damaged.

Diaphragmatic hernia A failure of the diaphragm to form completely, leaving a hole. Abdominal organs can protrude through the hole into the chest cavity and interfere with development of the heart and lungs. Usually life-threatening and requires emergent surgery.

Down syndrome (Trisomy 21) The chromosomal abnormality characterized by an extra copy of chromosome 21. In rare cases this syndrome is caused by *translocation.* The extra copy can be free-lying, or can be attached to some other chromosome, most frequently number 14. Down syndrome can occur in *mosaic,* so that there is a population of normal cells and a population of trisomy 21 cells. Down syndrome is characterized by moderate to severe mental retardation, sloping forehead, small ear canals, flat bridged nose and short fingers and toes. One third of infants have congenital heart disease, and one third have duodenal atresia. (Both can be present in the same infant.) Affected people can survive to middle or old age. There is an increased incidence of Alzheimer disease in adults with Down syndrome.

Ebstein anomaly A congenital heart defect in which the tricuspid valve is displaced downward into the right ventricle causing abnormal patterns of cardiac circulation.

Edwards syndrome (Trisomy 18) The chromosomal abnormality characterized by an extra copy of chromosome 18. The extra chromosome can be free lying or attached to another chromosome. Trisomy 18 can occur in mosaic. Edwards syndrome is characterized by

mental retardation, neonatal hepatitis, low-set ears, skull malformation and short digits. Cardiac and renal anomalies are also common. Survival for more than a few months is rare.

Encephalocele The protrusion of the brain substance through a defect in the skull.

Endocardial cushion defect A variety of septal defects (malformations of the walls separating the two atria and two ventricles of the heart) resulting from imperfect fusion of the endocardial cushions in the embryonic heart.

Epispadias A congenital defect in which the urinary meatus (urinary outlet) opens above (dorsal to) the normal position. The urinary sphincters are defective, so incontinence does occur. Surgical correction is aimed at correcting incontinence and permitting sexual functioning. The corresponding defect in females is rare. *See also Hypospadias.*

Esophageal stenosis or atresia A narrowing or incomplete formation of the esophagus. Usually a surgical emergency. Frequently associated with a tracheoesophageal fistula.

Fetal alcohol syndrome A constellation of physical abnormalities (including characteristic abnormal facial features and growth retardation), and problems of behavior and cognition in children born to mothers who drank alcohol during pregnancy.

Fistula An abnormal passage from an internal organ to the body surface or between two internal organs or structures.

Gastroschisis A congenital opening of the abdominal wall with protrusion of the intestines. This condition is surgically treated. Contrast with Omphalocele, below.

Hirschsprung disease The congenital absence of autonomic ganglia (nerves controlling involuntary and reflexive movement) in the muscles of the colon. This results in immobility of the intestines and may cause obstruction or stretching of the intestines. This condition is repaired surgically in early childhood by the removal of the affected portion of the intestine.

Holoprosencephaly Failure of the brain to develop into two equal halves, so there is structural abnormality of the brain. There may be associated midline facial defects including cyclopia (fusion of the eye orbits into a single cavity containing one eye) in severe cases. About half the cases are probably due to a single gene defect (the HPE gene). Frequently occurs with Trisomy 13.

Hydrocephaly The abnormal accumulation of fluid within the spaces of the brain.

Hyperplasia Overgrowth characterize by an increase in the number of cells of a tissue.

Hypoplasia A condition of arrested development in which an organ or part remains below the normal size or in an immature state.

Hypoplastic left heart syndrome Atresia, or marked hypoplasia, of the aortic opening or valve, with hypoplasia of the ascending aorta and defective development of the left ventricle (with mitral valve atresia). This condition can be surgically repaired in a series of three procedures over a period of one year. Transplantation is also a treatment. This condition is usually fatal in the first month of life if not treated.

Hypospadias A congenital defect in which the urinary meatus (urinary outlet) is on the underside of the penis or on the perineum (area between the genitals and the anus). The urinary sphincters are not defective so incontinence does not occur. The condition may be surgically corrected if needed for cosmetic, urologic, or reproductive reasons. The corresponding defect in women is rare. *See also epispadias*

Limb defects *See Reduction defects.*

Meninges Membranes that cover the brain and spinal cord.

Microcephaly The congenital smallness of the head, with corresponding smallness of the brain.

Microphthalmia The congenital abnormal smallness of one or both eyes. Can occur in the presence of other ocular defects.

Microtia A small or maldeveloped external ear and atretic or stenotic external auditory canal.

Mosaic In genetics, this refers to an individual organism that has two or more kinds of genetically different cell types. The degree of abnormality depends on the type of tissue containing affected cells. Individuals may vary from near normal to full manifestation of the genetic syndrome. Can occur in any chromosome abnormality syndrome.

Neural tube defect A defect resulting from failure of the neural tube to close in the first month of pregnancy. The major conditions include anencephaly, spina bifida, and encephalocele.

Obstructive genitourinary defect Stenosis or atresia of the urinary tract at any level. Severity of the defect depends largely upon the level of the obstruction. Urine accumulates behind the obstruction and damages the organs.

Omphalocele The protrusion of an organ into the umbilicus. The defect is usually closed surgically soon after birth. Contrast with Gastroschisis.

Patau syndrome (Trisomy 13) The chromosomal abnormality caused by a extra chromosome 13. The extra copy can be free-lying, or can be attached to some other chromosome. Patau syndrome can occur in *mosaic* so that there is a population of normal cells and a population of trisomy 13 cells. Patau syndrome is characterized by impaired midline facial development, cleft lip and palate, polydactyly and mental retardation. Most infants do not survive beyond 6 months of life.

Patent ductus arteriosus A blood vessel between the pulmonary artery and the aorta. This is normal in fetal life, but can cause problems after birth, particularly in premature infants. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. The vast majority close spontaneously and cause no problems. Medical or surgical correction may be done. This is only an abnormality if it causes significant medical problems.

Pulmonary artery anomaly Abnormality in the formation of the pulmonary artery such as stenosis or atresia. See also common truncus.

Pulmonary valve atresia or stenosis A congenital heart condition characterized by absence or constriction of the pulmonary valve. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can vary from mild to severe. Mild forms are relatively well tolerated and require no intervention. More severe forms are surgically corrected.

Pyloric stenosis A narrowing of the pyloric sphincter at the outlet of the stomach. This causes a blockage of food from the stomach into the small intestine. Usually treated surgically.

Reduction defects of the lower limbs The congenital absence of a portion of the lower limb. There are two general types of defect, transverse and longitudinal. Transverse defects appear like amputations, or like missing segments of the limb. Longitudinal defects are missing rays of the limb (for example, a missing tibia and great toe).

Reduction defects of the upper limbs The congenital absence of a portion of the upper limb. There are two general types of defect, transverse and longitudinal. Transverse defects appear like amputations, or like missing segments of the limb. Longitudinal defects are missing rays of the limb (for example, a missing radius and thumb).

Renal agenesis or dysgenesis The failure, or deviation, of embryonic development of the kidney.

Spina bifida A neural tube defect resulting from failure of the spinal neural tube to close. The spinal cord and/or meninges may or may not protrude. This usually results in damage to the spinal cord with paralysis of the involved limbs. Includes myelomeningocele (involving both spinal cord and meninges) and meningocele (involving just the meninges).

Stenosis A narrowing or constriction of the diameter of a bodily passage or orifice.

Stenosis or atresia of large intestine, rectum and anus The absence, closure or constriction of the large intestine, rectum or anus. Can be surgically corrected or bypassed.

Stenosis or atresia of the small intestine A narrowing or incomplete formation of the small intestine obstructing movement of food through the digestive tract.

Tetralogy of Fallot A congenital cardiac anomaly consisting of four defects: ventricular septal defect, pulmonary valve stenosis or atresia, displacement of the aorta to the right, and hypertrophy of right ventricle. The condition is corrected surgically.

Tracheoesophageal fistula An abnormal passage between the esophagus and trachea. Leads to pneumonia. Corrected surgically. It is frequently associated with esophageal atresia.

Translocation The rearrangement of genetic material within the same chromosome or the transfer of a segment of one chromosome to another one. People with balanced translocations do not always manifest genetic syndromes, but may be carriers of genetic syndromes and can have children with unbalanced translocations. Can occur with any chromosomal anomaly syndrome.

Transposition of the great vessels A congenital malformation in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle (opposite of normal), so that the venous return from the peripheral circulation is recirculated without being oxygenated in the lungs. Immediate surgical correction is needed. When this is not associated with other cardiac defects, and not corrected, it is fatal.

Tricuspid valve atresia or stenosis A congenital cardiac condition characterized by the absence or constriction of the tricuspid valve. The opening between the right atrium and right ventricle is absent or restricted, and normal circulation is not possible. This condition is often associated with other cardiac defects. This condition is surgically corrected depending on the severity.

Trisomy A chromosomal abnormality characterized by one more than the normal number of chromosomes. Normally, cells contain two of each chromosome. In trisomy, cells contain three copies of a specific chromosome.

Trisomy 13 *See Patau Syndrome.*

Trisomy 18 *See Edwards Syndrome*.

Trisomy 21 *See Down Syndrome*.

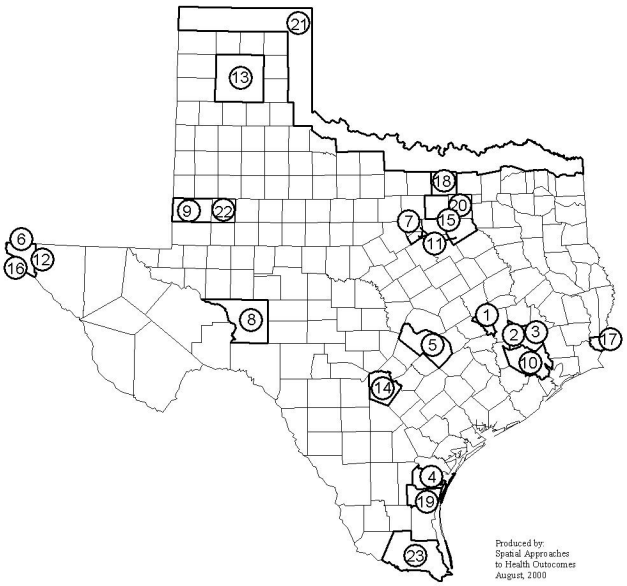
Truncus arteriosus *See Common truncus*.

Ventricular septal defect (VSD) A congenital cardiac malformation in which there are one or several openings in the ventricular septum (muscular and fibrous wall between the right and left ventricle or right and left lower chambers of the heart) allowing a mixing of oxygenated and unoxygenated blood. The openings vary in size and may resolve without treatment or require surgical treatment.

Appendix D: Cluster Investigations–1996 & 1997

In addition to routine data collection, we conduct investigations of birth defects clusters throughout the state. Health care professionals and the public can report apparently unusual concentrations of birth defects to TBDMD. Birth defect investigations are then initiated to determine if these clusters represent statistically significant variation. In 1996 and 1997, 23 clusters were investigated, as indicated below.

Figure 17: Location of Cluster Investigations, 1996-1997



	Condition	Area
1.	Anencephaly	Brazos (Bryan/College Station)
2.	Anencephaly	Montgomery (Conroe)
3.	Anencephaly	Montgomery (The Woodlands)
4.	Anencephaly	Nueces
5.	Anopthalmia	Travis and Bastrop
6.	Biliary atresia	El Paso
7.	Chromosomal defects	Hood
8.	Cleft lip and cleft palate	Crockett (Ozona)
9.	Cleft lip and cleft palate	Gaines and Dawson
10.	Down syndrome	Harris (Houston)
11.	Down syndrome	Hood, Somervell, and Ellis
12.	Heart defects	El Paso (Fabens)
13.	Multiple birth defects	Armstrong, Carson, Potter, and Randall
14.	Multiple birth defects	Bexar (Kelly AFB)
15.	Multiple birth defects	Dallas, Ellis, and Tarrant (Grand Prairie)
16.	Multiple birth defects	El Paso
17.	Multiple birth defects	Orange (Vidor)
18.	Multiple birth defects and conditions	Denton (Lewisville)
19.	Multiple birth defects and conditions	Kleberg (Kingsville)
20.	Neural tube defects	Dallas
21.	Renal agenesis	Texas/Oklahoma border
22.	Spina bifida	Dawson
23.	Ventricular septal defect	Cameron, Willacy, and Hidalgo

For background and response information, a detailed report of these investigations is available upon request from the Texas Birth Defects Monitoring Division, 512-458-7232 or http://www.tdh.state.tx.us/tbdmd/index.htm.

Appendix E: Research Using Data from the Texas Birth Defects Registry

Once in the Texas Birth Defects Registry, a case infant's family may be invited to participate in research studies. Epidemiologists and clinicians conduct epidemiological research to investigate the causes of birth defects. Members of the Texas Department of Health Institutional Review Board annually evaluate the protocols for each study to protect the privacy and other concerns of participants.

Below are descriptions of ongoing and completed research projects using data contained in the Texas Birth Defects Registry.

ONGOING RESEARCH USING REGISTRY DATA

Peter Langlois, Ph.D., Mark A. Canfield, Ph.D., and Angela Scheuerle, M.D., in collaboration with investigators in seven other states, serve as the co-principal investigators of the ***National Birth Defects Prevention Study*** (NBDPS). The on-going case-control study consists of conducting a one-hour computer-assisted telephone interview with 300 Texas women per year who delivered on or after October 1, 1997. Interviews are conducted with cultural sensitivity and can be done in either English or Spanish. Each family enrolled in the study will also be invited to donate swabs of cells from inside their cheeks. We will extract the DNA from their cells to study the genetic factors associated with birth defects. Among all eight states, principal investigators will enroll more than 16,000 women across the nation over five years. About 12,000 of the enrolled women will have had children or pregnancies affected by birth defects. An additional 4,000 women are mothers of infants with no birth defects.

Mark A. Canfield, Ph.D., and Kim Waller, Ph.D., are the principal co-investigators of the ***Study of Hispanic Origin, Maternal Obesity, and Disorders of the Central Nervous System***. They are examining the metabolic-genetic-environmental risks associated with certain birth defects of the brain and spinal cord. The study consists of a telephone interview of 300 mothers of Registry cases and controls delivered on or after October 1, 1997. Proposed date of completion is 2001.

Kate Hendricks, M.D., M.P.H., and Russell Larsen, Ph.D., are the principal co-investigators of the ***Texas Neural Tube Defect Surveillance and Intervention Project*** (partly funded by the Texas Birth Defects Research Center). The study, conducted since 1993, uses novel approaches to find causes of neural tube defects (NTDs), including anencephaly, spina bifida, and encephalocele. One approach uses Geographic Information Systems (GIS) to construct residential maps of affected and unaffected infants to study the proximity and exposure to pesticides and toxic sites. Another approach examines biological specimens from families to discover genetic and environmental factors related to NTDs. The proposed completion date is 2000.

Mark A. Canfield, Ph.D., Celia Kaye, M.D., Ph.D., and Maricella Aguilar, M.P.H., are the principal investigators of the ***Neural Tube Defect Recurrence Prevention Project***. This project is a statewide educational intervention for families with pregnancies affected by neural tube defects delivered in 1999, 2000, and 2001. Its purpose is to promote to mothers and health care providers the pre-conception consumption of high-dose folic acid to reduce the mother's risk of having subsequent NTD-affected pregnancies. The proposed completion date is 2001.

Mark A. Canfield, Ph.D., Wendy Nembhart, Ph.D., M.P.H., and Kim Waller, Ph.D., are the co-principal investigators for the study of ***Mortality Patterns Among Cases in the Texas Birth Defects Registry***. This study evaluates the patterns of survival of infants with different types of birth defects among cases delivered in 1996 and 1997. The proposed completion date is 2000.

John Belmont, M.D. Ph.D., is the principle investigator on ***Molecular Genetics of Congenital Left Ventricular Outflow Tract Obstruction***. The long term goal of this study is to identify genes contributing to the susceptibility to one pathophysiologic class of congenital heart defects. Specifically, the study will analyze congenital heart defects thought to result from flow abnormalities in

the left ventricular outflow tract (LVOT). The diagnoses of aortic valve stenosis, coarctation of the aorta, and hypoplastic left heart syndrome are included. This protocol will establish a bank of lymphoblastoid cell lines and derived DNA samples from patients, parents, and related family members affected with LVOT abnormalities. These samples will be used in genetic mapping and mutation studies that may ultimately clarify the origin of these defects. Identification of the susceptibility genes may contribute to the development of preventive strategies aimed at reducing the incidence of congenital heart defects.

COMPLETED RESEARCH USING REGISTRY DATA

Frequency of Prenatal Diagnosis of Birth Defects in Houston, Galveston and the Lower Rio Grande Valley, Texas 1995.

DK Waller, Ph.D., MA Pujazon, M.P.H., MA Canfield, Ph.D., AE Scheuerle, M.D., JLB Byrne, M.D. The University of Texas Houston Health Science Center School of Public Health and the Texas Birth Defects Monitoring Division, Bureau of Epidemiology, Texas Department of Health.

Background: Estimates of the proportion of birth defects diagnosed before birth exist for only a few types of birth defects and for a few geographic regions in the United States. This population based study examines rates of prenatal diagnosis for previously unstudied birth defects in a new geographic region.

Methods: Active surveillance of 23 categories of birth defects among 111,902 infants born in 77 birthing hospitals in Texas in 1995 identified 852 infants or fetuses with major birth defects. Surveillance was conducted by the Texas Birth Defects Monitoring Division of the Texas Department of Health. Two regions were covered: the Houston/Galveston metropolitan area and the Lower Rio Grande Valley of Texas. Rates of prenatal diagnosis were evaluated for 23 different types of birth defects, using proportions and 95 percent confidence intervals.

Results: One third of the 852 infants or fetuses with birth defects were prenatally diagnosed. Diagnosis rates varied greatly depending on the type of birth defect and were lower among infants born to black and Hispanic women. More than 60 percent of anencephaly, encephalocele, gastroschisis, and trisomies 13 and 18 were diagnosed antenatally. Most of the fetuses that were electively terminated had birth defects or combinations of birth defects that were potentially lethal. Prevalence rates for birth defects generally do not include fetuses that die or are electively terminated before 20 weeks of gestation. Thus, 36 percent of anencephaly, 21 percent of omphalocele, 15 percent of encephalocele, and between 7 and 10 percent of spina bifida, hydrocephaly, renal agenesis, and trisomies 13, 18, and 21 were not included in our published rates.

Conclusions: Published rates for specific types of birth defects are spuriously low. This should be considered when investigating alleged clusters and comparing rates of birth defects across geographic areas.

This study was completed in 1998. A manuscript describing these findings has been conditionally accepted by *Fetal Diagnosis and Therapy* for publication in 2000.

The Association Between Macrosomia and Major Congenital Birth Defects

DK Waller, Ph.D., MA Canfield, Ph.D., AE Schereule, M.D., A Keddie. The University of Texas Houston Health Science Center School of Public Health and the Texas Birth Defects Monitoring Division, Bureau of Epidemiology, Texas Department of Health.

Background: This study was undertaken to determine whether or not there is an excess of major congenital malformations among macrosomic infants. Macrosomia is defined as weighing more than 4000 grams at birth. Macrosomic infants are more likely to occur among diabetic women, obese women, and women who develop gestational diabetes during their pregnancy. Since these sub-groups of women are also known to have a higher risk of having infants with major malformations we

hypothesized that macrosomic infants may be more likely to have major congenital malformations.

Methods: The case-control study was undertaken using cases of congenital anomalies that were ascertained by the Texas Birth Defects Monitoring Division, an active population based surveillance system. Controls were obtained from Texas birth certificates. All cases and controls were delivered between January 1, 1995 and December 31, 1997. We included live born infants in the study. We excluded multiple births from the study because twins and higher order multiple births have significantly lower mean birth weights than singletons.

Results: We found an increased risk of all major congenital anomalies among macrosomic infants compared with the referent group of infants weighing 3500 to 3749 grams. The increased risk was present for all sub-groups of ethnicity and infant gender, except white males. The congenital anomalies that were increased among macrosomic infants include the following categories: encephalocele, holoprosencephaly, anomalies of the corpus collosum, anomalies of cardiac septal closure, all other congenital heart anomalies, anomalies of the circulatory system exclusive of heart, preaxial polydactly, rib and sternum anomalies, and omphalocele.

Conclusion: This study had adequate statistical power to conclude that there is an excess of all major congenital anomalies among infants with macrosomia. However, some of the associations that we observed between particular anomalies and macrosomia are based on small numbers and should therefore be interpreted cautiously. As expected, we also observed the well established excess of major congenital anomalies among low birth weight infants (< 2500 gm) and very low birth weight infants (<1500 grams). A manuscript describing these findings is under preparation.

Evaluation of a System for Linking Birth Defects Registry Records and Vital Records

MB Forrester, B.S., MA Canfield, Ph.D. The Texas Birth Defects Monitoring Division, Bureau of Epidemiology, Texas Department of Health.

Background: The intent of this investigation was to evaluate a procedure for linking Texas Birth Defects Monitoring Division (TBDMD) registry records to Texas Bureau of Vital Statistics (BVS) birth and fetal death database records.

Methods: Using Microsoft Access, the researchers attempted to match TBDMD registry records to BVS records using six variables (infant's last name, infant's first name, infant's date of birth, mother's first name, mother's date of birth, birth facility code). If an exact match could not be made on all six variables, then fewer variables were used in the program, and the differences in the remaining discordant variables were evaluated to determine whether the TBDMD and BVS records were a match.

Results: The TBDMD was able to successfully link 96.8 percent of its registry records to BVS records. The linkage rate was higher for live births (99.3 percent) than for fetal deaths (80.1 percent) and elective terminations (39.2 percent). The concordance between variables of matched records varied by the type of variable. Birth facility code and infant's date of birth demonstrated the highest degree of concordance while infant's last name and infant's first name demonstrated the lowest degree of concordance.

Conclusion: The TBDMD is able to link to BVS records with a high degree of success. This will allow the TBDMD to use BVS as a potential source of data not easily or efficiently obtained through the TBDMD's other data sources.

The editors wish to acknowledge the tremendous dedication of
Texas Birth Defects Monitoring Division field staff in
the challenging job of collecting the information
which is represented in this report.

Many thanks to the Garcia Family--Abelardo,
Norma, and Diana, who was born in
December 1999 with Down Syndrome

For More Information

This report is a publication of the
Texas Birth Defects Monitoring Division
Bureau of Epidemiology
Texas Department of Health
1100 W. 49th Street
Austin, Texas 78756
512-458-7232
http://www.tdh.state.tx.us/tbdmd/index.htm

Pub. No. 58-10772
Published September 2000